

# Bundle of His electrograms in congenital corrected transposition of the great arteries

## *A study of two adult cases*

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*His bundle electrograms were recorded in 2 adult cases of congenital corrected transposition of the great arteries. Both patients had prolonged HV conduction and a His potential that was wide. These findings are different from those reported in congenital atrioventricular block without ventricular inversion wherein an abnormality within the AH period has been found. It is suggested that these differences reflect different sites of disturbed conduction. In congenital corrected transposition of the great vessels, the abnormality is distal to the atrioventricular node, whereas in congenital atrioventricular block without ventricular inversion it is generally proximal to the atrioventricular node.*

His bundle electrocardiography has been used to localize the site of conduction disturbance in atrioventricular block (Damato *et al.*, 1969; Narula *et al.*, 1970a). Atrioventricular conduction abnormalities have been found in more than one-third of reported cases of congenital corrected transposition of the great arteries (Friedberg and Nadas, 1970). To determine the site of disturbance in 2 adult cases of congenital corrected transposition of the great arteries, His bundle electrocardiography was performed. The results of these studies are now reported and discussed in the light of previous morphological and electrophysiological studies.

### Methods

Patients were studied at the time of routine cardiac catheterization. Diazepam was administered one hour before catheterization and approximately 3 hours before His bundle electrograms were obtained. One patient (Case 2) had been receiving digoxin 0.25 mg each day; it was stopped 72 hours before study. No other drugs were administered during the preceding two weeks.

His bundle electrograms were recorded using the method described by Scherlag *et al.* (1969). Specifically, a multipolar catheter was advanced via a femoral vein across the right-sided atrioventricular (mitral) valve. His bundle electrograms and simultaneous surface electrocardiograms were recorded at a paper speed of 200 mm/sec on a multichannel recorder (Electronics for Medicine - DR-12).

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### Definition of terms

**PH:** interval from onset of P wave on the surface electrocardiogram to first high frequency component of His bundle electrogram (normal  $119 \pm 38$  msec) (Hecht *et al.*, 1973).

**HV:** interval from first high frequency component of His bundle electrogram to onset of QRS complex on surface electrocardiogram or V wave of ventriculogram, whichever occurred earlier (normal  $43 \pm 12$  msec) (Hecht *et al.*, 1973.)

**H potential:** high frequency deflection after isoelectric period following A wave (total duration was measured - normal 15-20 msec) (Narula *et al.*, 1970a).

**PR:** interval from onset of P wave to onset of QRS complex on surface electrocardiogram.

**Congenital corrected transposition of the great arteries:** a disorder of the heart in which the aorta arises from a left-sided anatomical right ventricle and the pulmonary artery arises from a right-sided anatomical left ventricle and wherein vena caval blood passes to the pulmonary artery and pulmonary venous blood to the aorta. Ventricular inversion is used interchangeably with congenital corrected transposition of the great arteries.

**Left ventricle ('RV'):** refers to the ventricle that has a fine trabecular pattern, a bicuspid atrioventricular valve, and does not have a crista supraventricularis (right-sided or venous ventricle).

**Right ventricle ('LV'):** refers to the ventricle that has a coarse trabecular pattern, a tricuspid atrioventricular valve, and a crista supraventricularis (left-sided or arterial ventricle).

## Case reports

### Case 1

A 19-year-old female college student was admitted to The Mount Sinai Hospital of New York for diagnostic cardiac catheterization. A heart murmur was first heard at 3 years of age. She had been asymptomatic and participated in competitive sports without difficulty. Because of uncertainty of diagnosis cardiac catheterization was performed.

Physical examination showed a thin, healthy woman. Venous and carotid pulse contours were normal. The first heart sound was normal. The second heart sound was single and heard best in the second left intercostal space. A grade 3/6 harsh ejection-type systolic murmur was heard at the base. Liver dullness was percussed on the right side. There was no cyanosis or clubbing.

The electrocardiogram showed absence of a Q wave in leads I and V6 and a QS complex in V1 and V2; the T wave was inverted in leads I and aVL and upright in V1-V6. The vectorcardiogram revealed leftward direction of initial forces of the QRS vector loop and anterior and rightward direction of the mean T vector (Fig. 1).

An angiogram of the left-sided ventricle, from which

the aorta arose, showed coarse trabeculation, an infundibulum, and crista supraventricularis (Fig. 2). The right-sided ventricle, from which the pulmonary artery arose, was smooth; there was, however, narrowing of the out-flow tract. Cardiac catheterization data are shown in Table 1.

### Case 2

A 37-year-old man, employed as a cook, was admitted to The Mount Sinai Hospital of New York for cardiac catheterization. At 22 years of age, he was rejected for military service because of 'heart trouble'. He experienced no difficulties until one month before admission to hospital, when he developed fever and cough. Despite symptomatic treatment for one week, he did not improve. He was found at this time to have a heart murmur and pulmonary congestion. He was treated with digoxin and diuretics with resolution of symptoms.

Physical examination showed a middle-aged man of healthy appearance. Blood pressure was 110/70 mmHg. Venous and carotid pulse contours were normal. There was a prominent left parasternal impulse. The first heart sound was soft; the second heart sound was single and was

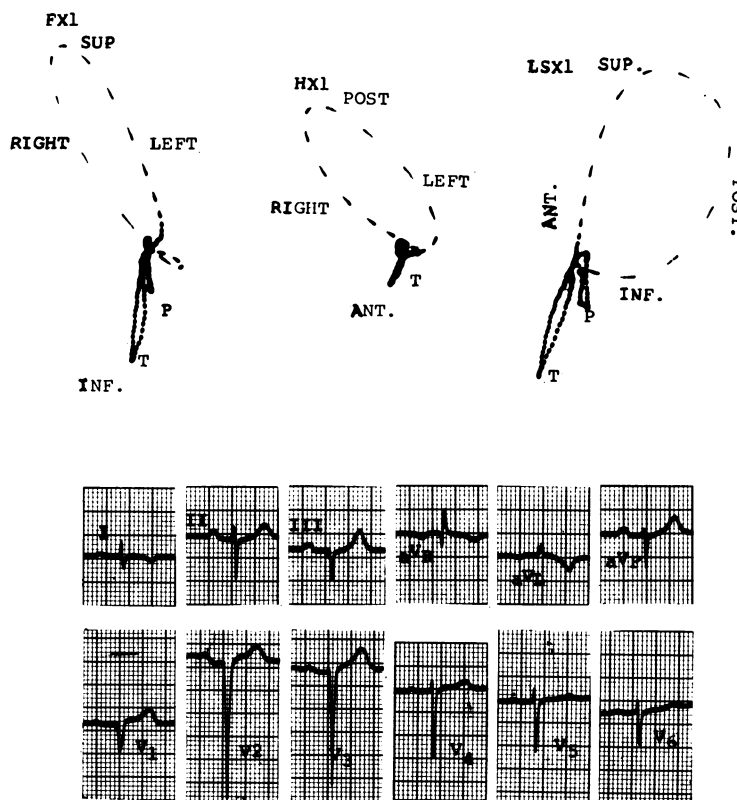


FIG. 1 Vectorcardiogram and electrocardiogram of Case 1. Electrocardiogram shows absence of Q wave in lead I and V6 and QS complex in V1 and V2. PR interval measures 0.20 sec. Vectorcardiogram shows leftward direction of initial forces of QRS vector loop.

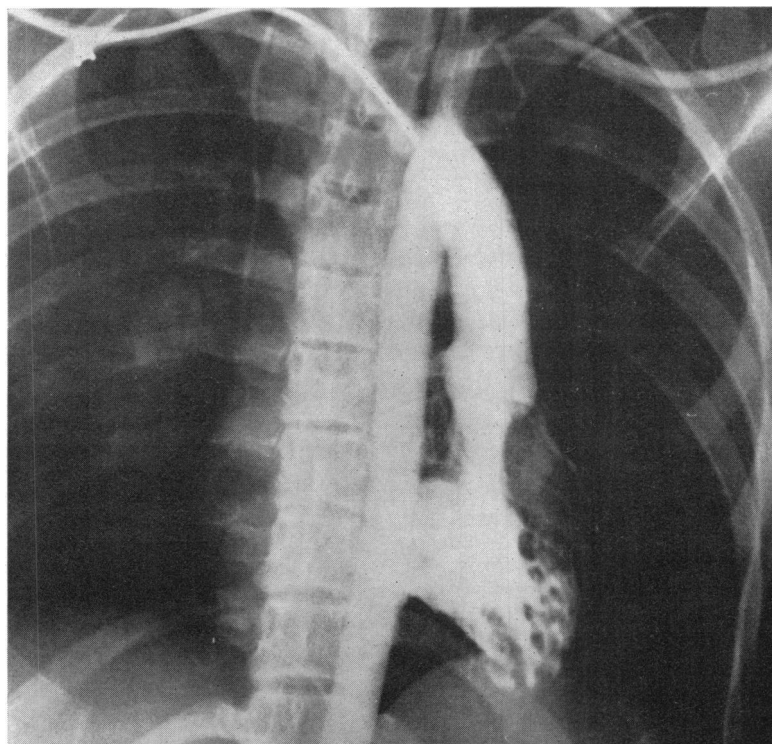


FIG. 2 Left-sided ventriculogram of Case 1 which demonstrates that the aorta arises from a coarsely trabeculated ventricle with an infundibulum.

TABLE 1 Results of cardiac catheterization

	Site	Pressures Dynamic	(mmHg) Mean	Oxygen saturation (%)
Case 1	IVC	—	—	80
	SVC	—	—	78
	RA	a = 6	4	80
	'RV'	46/3	—	80
	PA	—	—	—
	LA	—	6	94
	'LV'	99/5	—	—
Case 2	Ao	99/62	83	—
	IVC	—	—	79
	SVC	—	—	75
	RA	a = 14	6	75
		v = 10		
	'RV'	46/4	—	74
	PA	46/21	31	75
	PC	v = 31	17	—
	'LV'	130/9	—	—
	Ao	130/94	106	96

loudest in the second left intercostal space. A grade 3/6 high pitched pansystolic murmur was loudest along the left sternal border and radiated to the axilla, base, and back. Liver dullness was percussed on the right side. There was no cyanosis or clubbing.

The electrocardiogram showed first-degree atrio-ventricular block, absence of a Q wave in leads I and V6, and a T wave that was biphasic in leads I and aVL and upright in V1-V6. The vectorcardiogram revealed leftward direction of the initial forces of the QRS vector loop and anterior, inferior, and rightward direction of the mean T vector. The P vector loop was displaced posteriorly and the voltage of the maximum P vector was increased (Fig. 3).

An angiogram of the left-sided ventricle, from which the aorta arose, showed coarse trabeculation and an infundibulum (Fig. 4). The left atrium was enlarged, and the left-sided (tricuspid) atrioventricular valve was incompetent. Cardiac catheterization data are shown in Table 1.

## Results

The site of the conduction disturbance was found in both patients to be distal to the recorded His potential (Table 2; Fig. 5 and 6). Each P wave was

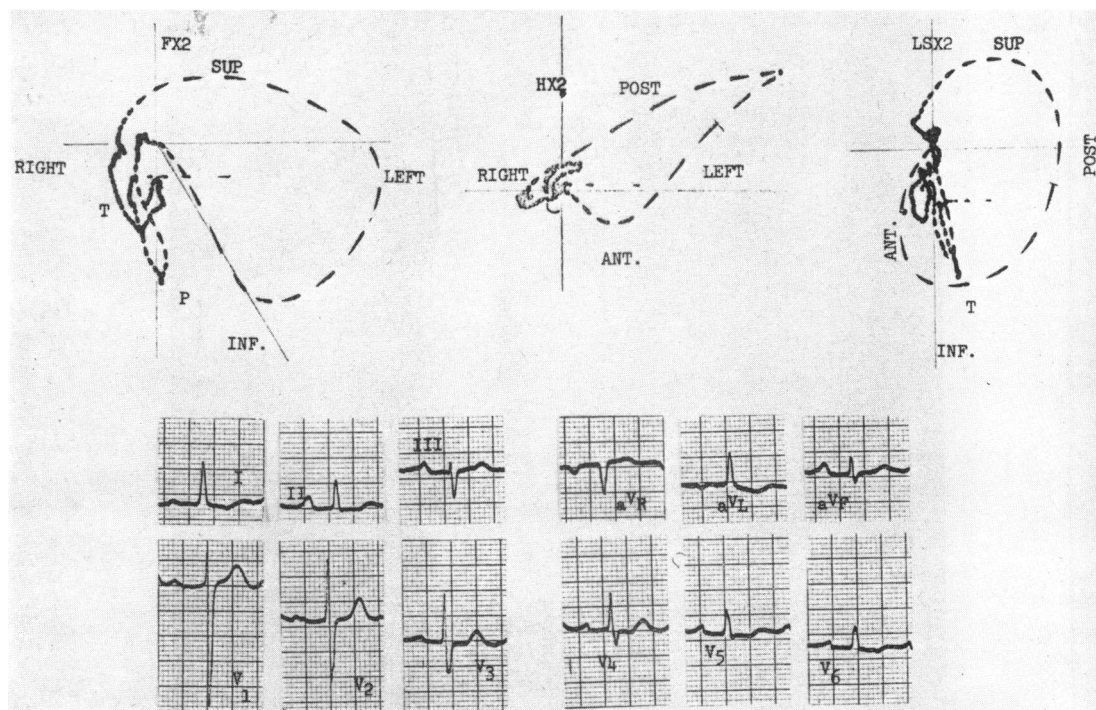


FIG. 3 Vectorcardiogram and electrocardiogram of Case 2. The electrocardiogram shows absence of a Q wave in lead I and V6. The PR interval measures 0.26 sec. The vectorcardiogram shows leftward direction of initial forces of QRS vector loop.

TABLE 2 Data obtained from bundle of His electrograms

	PR interval (msec)	PH time (msec)	HV time (msec)	H deflection (msec)	QRS duration (msec)
Case 1	190	122	68	25	70
Case 2	263	131	132	22	80

followed by a QRS complex. The PH interval was normal. The conduction time from the His potential to ventricular activation (HV) was increased. The His potential was prolonged and had an abnormal configuration. In one patient (Case 1) it was found to be polyphasic (Fig. 5) and in the other (Case 2) it was slurred (Fig. 6).

A second deflection was recorded in one patient (Case 2) as the catheter was advanced into 'RV' (at the time in which the V wave was prominent and the A wave had low voltage) (Fig. 7). This potential (H') was monophasic and occurred 35 msec before the V wave and may represent left bundle-branch potential.

### Discussion

Congenital corrected transposition of the great arteries has been reported to occur in less than 10 per cent of cases of congenital complete heart block (Nadas and Fyler, 1972). In a review of 18 patients with congenital complete heart block, however, 8 were found to have ventricular inversion (Kangos, Griffiths, and Blumenthal, 1967). Conversely, between one-third (Friedberg and Nadas, 1970) and two-thirds (Schiebler *et al.*, 1961) of patients with congenital corrected transposition of the great arteries have an atrioventricular conduction disturbance. The most common abnormality is a prolonged PR interval (Anderson, Lillehei, and Lester,

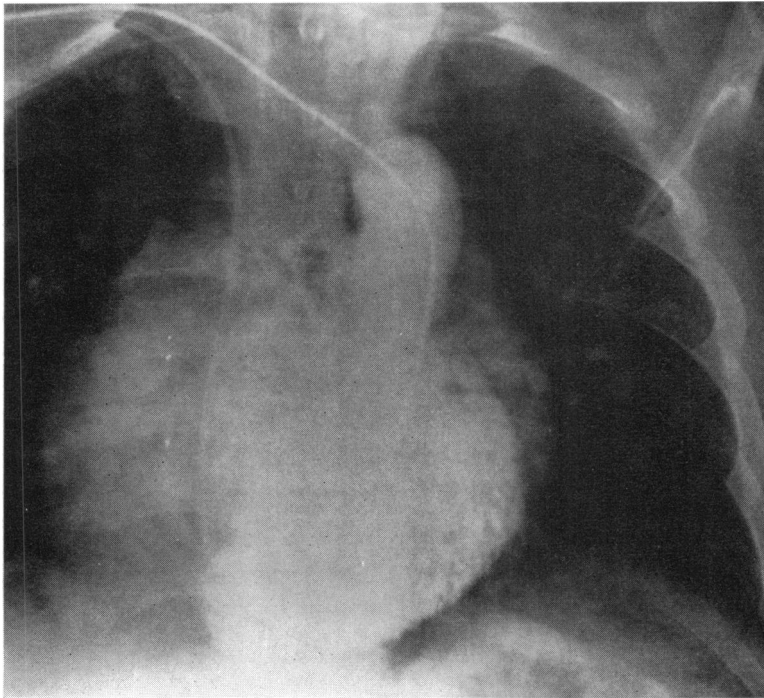


FIG. 4 Left-sided ventriculogram of Case 2 which demonstrates that the aorta arises from a coarsely trabeculated ventricle with an infundibulum.

1957; Friedberg and Nadas, 1970; Schiebler *et al.*, 1961). When complete heart block is found, the QRS complexes are narrow and the ventricular rate is usually greater than 40 beats/minute (Walker *et al.*, 1958; Kangos *et al.*, 1967).

In this study of 2 adult patients with congenital corrected transposition of the great arteries, His bundle electrograms demonstrated that the HV conduction time was prolonged whereas the AH interval was normal (Table 2; Fig. 5 and 6). Moreover, the His bundle potential was broad. In one patient (Case 2) it was slurred and in the other (Case 1) it was polyphasic. Prolonged HV conduction time indicates disease in the His-Purkinje system (Narula and Samet, 1970). Furthermore, increased HV conduction time in conjunction with narrow QRS complexes suggest that the abnormality is localized in the His bundle (Narula *et al.*, 1970b). An abnormal His bundle potential supports such an interpretation (Narula and Samet, 1970).

In the only previous report of His bundle electrograms in this disorder, Rosen *et al.* (1971) recorded a biphasic deflection that followed the P wave by 220 msec. A nonconducted P wave was recorded which was not followed by this deflection. Close

inspection of the published recordings suggests that an early, broad deflection may have been present. Labelled His bundle potentials may represent, therefore, either 'split His' (Narula and Samet, 1970) or left bundle-branch potential.

His bundle electrograms of complete heart block associated with congenital corrected transposition of the great arteries have not been recorded. In congenital complete heart block without ventricular inversion the site of the disturbed conduction has been localized proximal to the His potential (Kelly *et al.*, 1972). As one might expect from these His bundle electrograms, morphological studies of the conduction system in uncomplicated congenital complete heart block have shown that usually there is a discontinuity of the atrial musculature and the AV node (Lev *et al.*, 1971).

In congenital corrected transposition of the great arteries, however, the defect has been found to be distal to the AV node (Yater, 1929; Walmsley, 1931; Lev, Licata, and May, 1963; Anderson, Jones, and Arnold, 1971). In a case report by Yater (1929), complete separation of the AV node and His bundle was found. Walmsley (1931) attributed heart block to an abnormally long His bundle. Unlike the

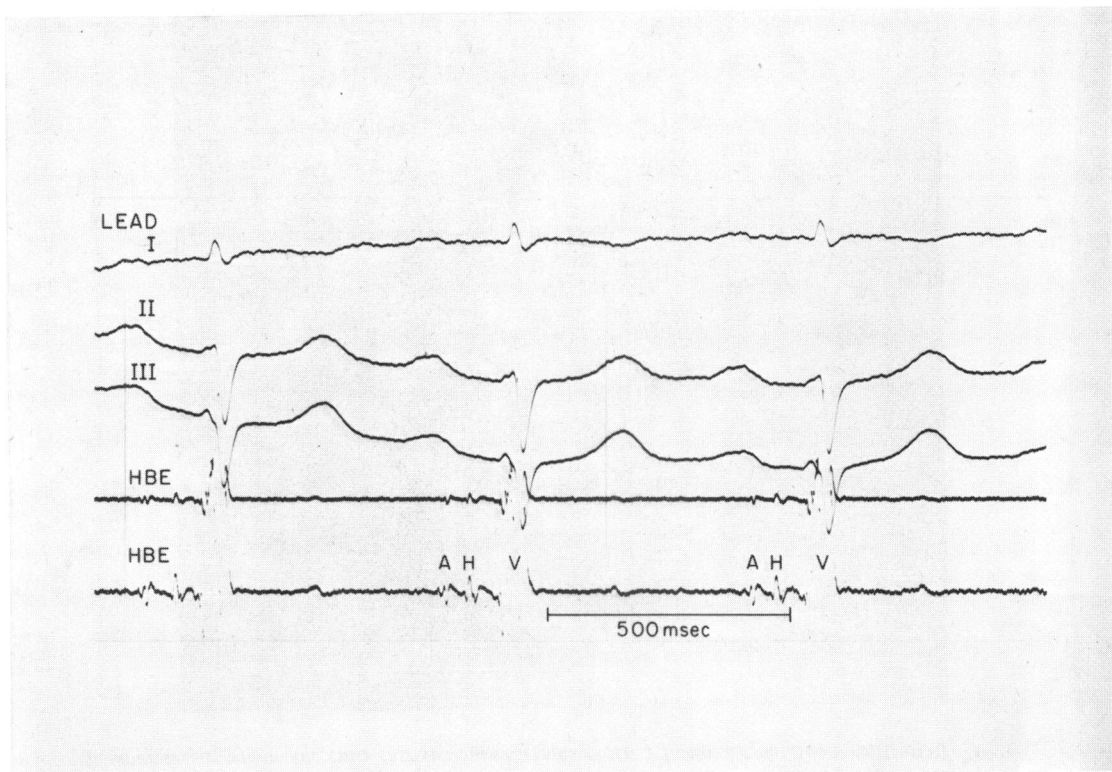


FIG. 5 His bundle electrograms of Case 1, which show a prolonged HV interval with a wide, polyphasic His potential.

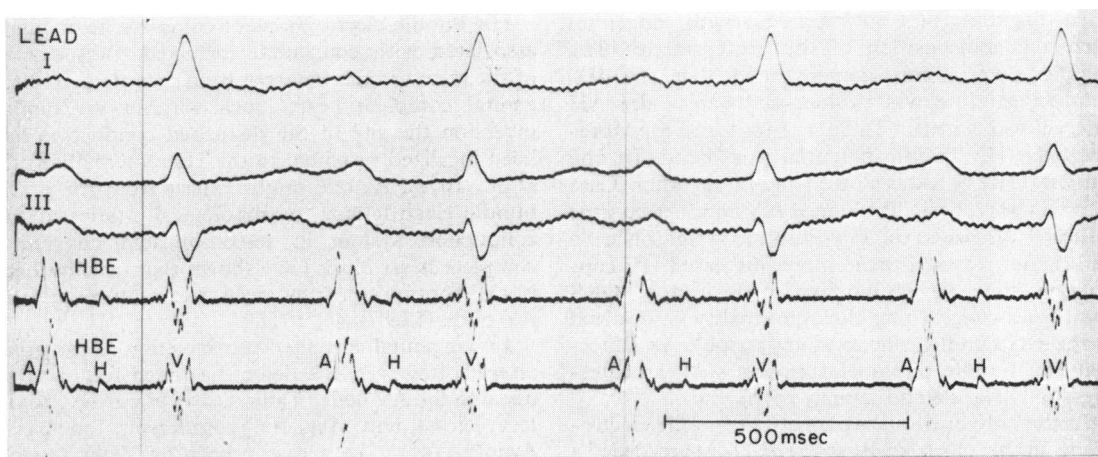


FIG. 6 His bundle electrograms of Case 2, which show a prolonged HV interval with a wide, slurred His potential.

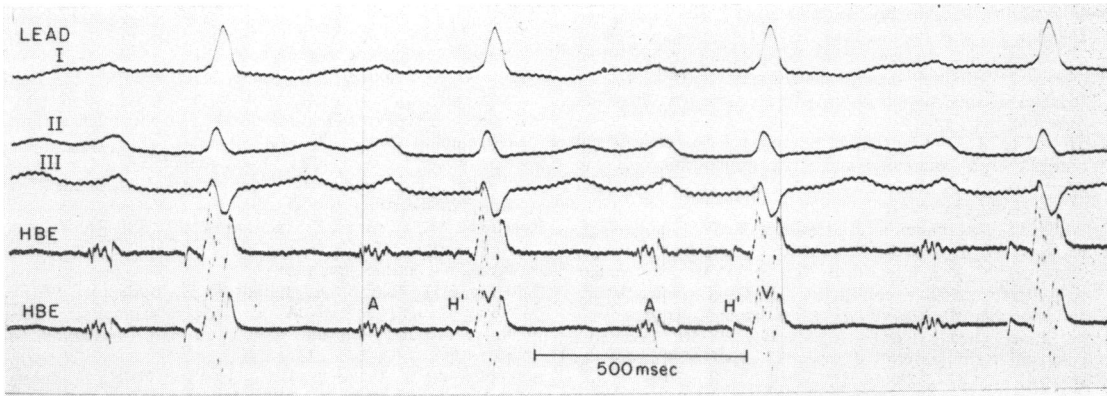


FIG. 7 Electrograms of Case 2 as the catheter was advanced into the right-sided ventricle (A wave low voltage and V wave prominent). A monophasic potential (H') preceding the V wave by 35 msec may represent left bundle-branch potential.

normal heart in which a part of the right atrium is contiguous with the left ventricle, in congenital corrected transposition of the great arteries, a portion of the left atrium and right-sided ventricle (anatomical left ventricle) are in continuity. The His bundle must, therefore, traverse an additional segment of the left atrium before reaching the ventricle. Walmsley (1931) observed, also, fibrosis in the His bundle. Lev *et al.* (1963) found that the His bundle and its branches were inverted in a mirror image of normal. Inversion of the AV node was also noted; however, anomalous coronary sinus drainage into the left atrium was present, perhaps accounting for this finding (Patten, 1956). In 3 patients, Anderson *et al.* (1971) found that the AV node and His bundle were separated and that the His bundle was either poorly developed or absent. These investigators found, however, that in one patient whose electrocardiogram showed a short PR interval, poor development of the approaches to the AV node was present whereas the AV node and His bundle were normal.

Two embryological observations appear to account for the frequent occurrence of conduction disturbance in this disorder: 1) discordance of the atrial situs and bulbo-ventricular loop (De la Cruz *et al.*, 1959); 2) development of the AV node with the atrium and the His bundle with the ventricle; proximal AV node apparently arises from the left horn of sinus venosus (Patten, 1956; James, 1970; Anderson and Taylor, 1972) whereas the His bundle arises from the AV canal musculature, a segment of which forms the distal AV node (Anderson and Taylor, 1972). Failure of the AV node and His bundle to join would, therefore, not be unexpected

(James, 1970) and would account for either delayed HV conduction time – perhaps, related to slow conduction via non His-Purkinje tissue – or complete heart block.

His bundle electrograms would be of interest in congenital corrected transposition of the great arteries with complete heart block. In view of the morphological studies reviewed herein and the His bundle recordings reported on, it is suggested that His electrograms of complete heart block associated with congenital corrected transposition of the great arteries may show any of the previously reported heart block patterns: paired A wave and His deflection dissociated from the V wave; paired His deflection and V wave dissociated from the A wave; and, perhaps, split His deflections, a proximal deflection paired to an A wave and a distal paired to the V wave (Narula *et al.*, 1970b; Narula and Samet, 1970).

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